

Right-sided subcutaneous implantable cardioverter-defibrillator placement in a patient with dextrocardia, tetralogy of Fallot, and conduction disease



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Introduction

Patients with congenital heart disease may be ideal candidates for completely subcutaneous implantable cardioverter-defibrillator (S-ICD) systems because of the presence of intracardiac shunts, limited venous access, and intracardiac anatomy that may be contraindicated to transvenous implantable cardioverter-defibrillator (ICD) systems. However, even S-ICD systems may require nonstandard placement because of anatomic abnormalities in patients with complex congenital heart disease. We describe the placement of an S-ICD on the right side of the thorax in a patient with tetralogy of Fallot and dextrocardia and discuss the use of S-ICDs in the pediatric and adult congenital heart disease populations.

Case report

The patient was a 21-year-old man with a history of dextrocardia, tetralogy of Fallot, and Klinefelter syndrome. His history was significant for multiple cardiac surgeries and sternotomies, including placement of a right modified BT shunt in the newborn period, complete repair of tetralogy of Fallot at age 1 year (ventricular septal defect closure, right ventricular outflow tract [RVOT] patch augmentation, resection of infundibular tissue in the RVOT), pulmonary valve replacement with intraoperative cryoablation of the right ventricle (RV)/RVOT due to ventricular tachycardia,

tricuspid valve annuloplasty, and modified right atrial maze procedure because of recurrent atrial flutter. Due to a history of recurrent ventricular tachycardia and easily inducible ventricular fibrillation (VF) on a prior ventricular stimulation study in the electrophysiology laboratory, he underwent placement of a left-sided transvenous dual-chamber dual-coil ICD at age 14 years. Defibrillation safety testing at that time failed, and the patient underwent placement of a subcutaneous coil with resultant successful safety testing. He subsequently developed severe pulmonary and tricuspid regurgitation and most recently underwent repeat pulmonary valve replacement (33-mm bioprosthetic), tricuspid valve replacement (31-mm bioprosthetic), RVOT patch augmentation, and planned removal of the transvenous ICD along with placement of an epicardial dual-chamber ICD. During this surgery, the sternum could not be fully opened because of adhesion of the RVOT to the sternum. Therefore, he was cannulated for bypass in the right femoral vessels. The pulmonary and tricuspid valves were replaced without incident, and the intracardiac portion of the transvenous ICD system was partially removed. Before the epicardial system could be completely implanted, however, there was difficulty maintaining bypass flow, and the abdomen was found to be distended. The sternotomy was extended inferiorly to a full laparotomy to relieve abdominal compartment syndrome, and the patient was returned to the intensive care unit with an open abdomen and chest. The abdomen and chest were closed 3 days later, but the epicardial ICD system was not placed at that time because of the patient's recurrent fevers and the concern for infection.

After the patient's full surgical recovery there were detailed discussions with the family and care team regarding options for ICD placement. Because of the risks associated with a repeat sternotomy and the concerns with placing a new transvenous lead across the patient's new prosthetic tricuspid valve, we elected to place a right-sided S-ICD. Considering the patient's history of atrial flutter, which was responsive to antitachycardia pacing

KEYWORDS Subcutaneous implantable cardioverter-defibrillator; Congenital heart disease; Pediatrics; Tetralogy of Fallot; Dextrocardia

ABBREVIATIONS ATP = antitachycardia pacing; ICD = implantable cardioverter-defibrillator; RV = right ventricle; RVOT = right ventricular outflow tract; S-ICD = subcutaneous implantable cardioverter-defibrillator; VF = ventricular fibrillation (Heart Rhythm Case Reports 2015;1:186–189)

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KEY TEACHING POINTS

- Patients with congenital heart disease may be ideal candidates for completely subcutaneous implantable cardioverter-defibrillator (S-ICD) systems because of the presence of intracardiac shunts, limited venous access, and intracardiac anatomy that may be contraindicated to transvenous ICD systems.
- S-ICDs can be placed in nonstandard locations (eg, right thorax) and can be effective in children and in patients with complex congenital heart disease.
- S-ICDs can be used successfully in combination with standard transvenous systems having antitachycardia pacing capabilities in patients with concomitant atrial arrhythmias.
- Careful screening, especially in patients with conduction disease, in different positions and with exercise is important before implantation.

(ATP), the decision was made to leave the transvenous atrial lead in place connected to the prior transvenous ICD

generator to be used for future potential therapy for atrial flutter.

Detailed screening for the S-ICD was performed before device placement. Baseline 12-lead ECG demonstrated sinus rhythm with right bundle branch block and QRS duration of 200 ms. Initial ECG vector screening revealed that the patient would be an adequate candidate based on screening with the generator in the right axilla and the coil placed on the right of the sternum. Screening with the generator in the left axilla and the coil to the left of the sternum, as well as screening with the generator in the right axilla and the coil to the left of the sternum, failed to meet adequate sensing thresholds (Figure 1). Because of concern about the patient's risk for inappropriate shocks due to his history of atrial flutter and wide QRS duration, screening was also performed on the treadmill and he passed the screening at high heart rates.¹

A Boston Scientific SQ-RX S-ICD (Boston Scientific, Marlborough, MA) was placed with the generator in the right axilla, the inferior portion of the coil to the right of the xyphoid process and the tip of the coil to the immediate right of the sternum with careful attention to avoid having the tip of the coil touch a sternal wire. Sensing through the device was noted to be best in the alternate sensing mode (from inferior portion of the coil to the generator). Sensing through the S-ICD was also checked with atrial pacing, rapid atrial pacing, and high-output atrial pacing

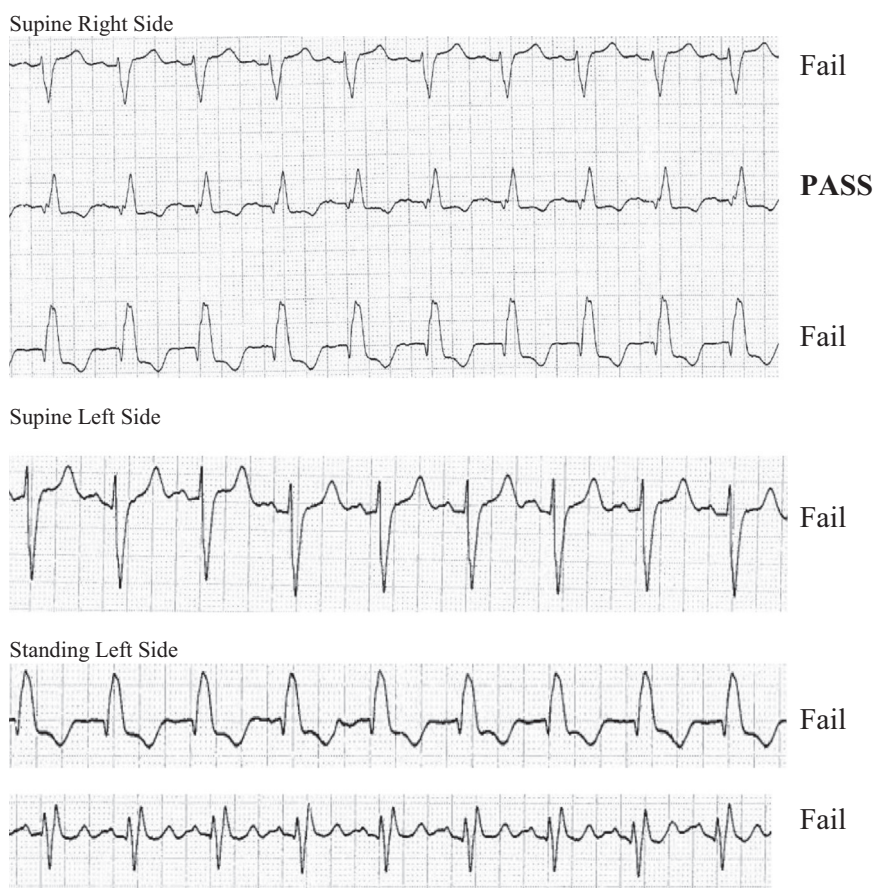


Figure 1 Representative screening electrograms through the programmer before device implantation. Although all screening electrograms on the left thorax failed screening, 1 lead configuration on the right side of the thorax passed the screening algorithm supine, standing, and with exercise.

from the transvenous atrial lead to ensure there were no problems with oversensing. Safety testing was performed in the electrophysiology laboratory at the time of implantation. A 50-Hz burst induced VF with adequate sensing and termination of VF with restoration of sinus rhythm after 18 seconds from one 65-J shock. His previously implanted transvenous system was left in place, with only the transvenous atrial lead connected to the ICD generator and programmed AAI at 60 bpm with the potential for

future manual ATP because of his history of recurrent atrial flutter (Figure 2).

Discussion

The use of ICDs in children and young adults with congenital heart disease has evolved over the past several decades. Implantation of transvenous and epicardial systems in these patients is not without long-term risks. The incidence of lead fractures, inappropriate shocks, venous occlusion, and infection seems to be higher in this population than in adults and can be as high as 30%.²⁻⁴ In addition, many patients cannot receive transvenous ICD systems because of single-ventricle physiology, the presence of intracardiac shunts, small patient size, and limited venous access to the heart.⁵ Thus, placement of an S-ICD may provide an excellent alternative to epicardial or transvenous systems in a select group of these patients.

In this report, we describe the use of an S-ICD system on the right side of the thorax in a young adult with a history of dextrocardia and congenital heart disease. The S-ICD system typically is placed in the left axilla, with the subcutaneous coil positioned to the left of the sternum.⁶ There are currently few reports in the literature on use of an S-ICD system in nonstandard positions.⁷⁻¹¹ In our patient, the S-ICD system functioned appropriately and effectively with positioning of the generator in the right axilla and the subcutaneous coil on the right side of the chest. Defibrillation safety testing also revealed excellent sensing during VF and an adequate defibrillation safety margin on the right side of the thorax.

There are also few reports of placement of S-ICD systems in children.^{12,13} Griksaitis et al¹⁴ reported on S-ICD use in 23 children, and Pettit et al¹³ reported on its use in 9 children (all on the left side of the thorax), with no difference in rates of pocket hematoma, infection, or inappropriate shocks compared to standard transvenous systems. Use of an S-ICD in a young patient who requires ICD placement is appealing because it avoids the need for transvenous leads, thus potentially averting venous occlusion and the notable risk of lead extraction in the event of lead failure or fracture.¹⁵ Although the S-ICD cannot be used as sole therapy in a patient requiring chronic pacing or ATP, we demonstrated in this case that the S-ICD system can be used in conjunction with a previously placed transvenous system that may still provide ATP if needed. Thus, the S-ICD can be used effectively in the right thorax in patients with complex congenital heart disease and dextrocardia.

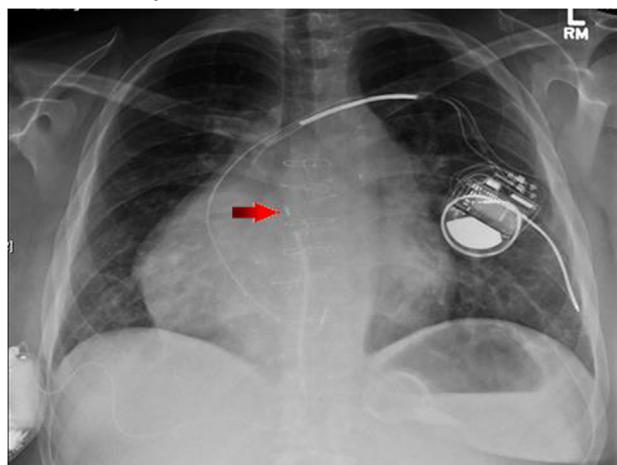
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A PA Projection



B Lateral projection



Figure 2 Chest radiograph after subcutaneous implantable cardioverter-defibrillator (S-ICD) placement. Posteroanterior (A) and lateral (B) chest x-ray films in the patient with dextrocardia and tetralogy of Fallot after S-ICD placement. Note the S-ICD generator in the right axilla and defibrillation coil immediately to the right of the sternum (red arrow). A transvenous atrial lead connected to the transvenous ICD system in the left delto-pectoral area was left in place to provide atrial antitachycardia pacing (ATP) because of the patient's history of atrial flutter that was responsive to ATP.

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